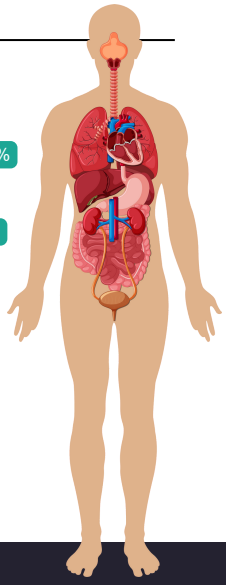
Neuroendocrine Cancer UK
Incorporating The Ann Edgar Trust

NEUROENDOCRINE CANCER

An overview for Healthcare Professionals

KEY FACTS

- **The incidence of neuroendocrine cancer has increased 371% (1995-2018), and it is now the 10th most prevalent cancer in England and the second most prevalent GI cancer.(1)**
- It is a cancer that starts in the neuroendocrine cells scattered throughout the body. Like nerve cells, neuroendocrine cells receive messages; like endocrine cells, they can release hormones. Therefore, a diagnosis of neuroendocrine cancer can lead to symptoms both as a consequence of the tumour site and hormone (hyper)secretion.
- **Neuroendocrine cancer is challenging to diagnose** as it can occur almost anywhere in the body, can be asymptomatic, and early symptoms can mimic more common pathologies such as IBS, asthma, menopause, and anxiety.(2)
- **Neuroendocrine cancer has a huge impact on patients' Quality of Life** (and their families) due to cancer and hormonal symptoms, relative rarity of diagnosis, information and diagnostic barriers and delays, alongside an often incurable, uncertain prognosis.(3)
- More than 50% of all cases will have advanced disease (Stage III & IV) at the time of diagnosis.(4)
- **Diagnosing patients earlier is life-changing because there are treatments that can improve the prognosis as well as symptoms even where metastatic (spread of) cancer exists.**



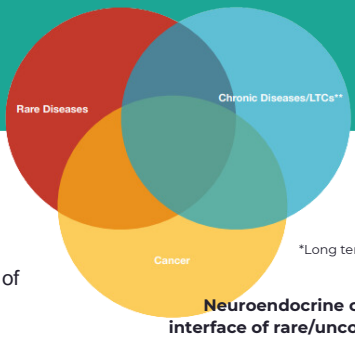
Neuroendocrine cancers may develop almost anywhere in the body, most commonly within the respiratory or digestive tracts (GI tract /pancreas).

The likely locations of neuroendocrine cancer. (*5*)

"Any cancer diagnosis is devastating, then you have the added confusion and bewilderment of dealing with a rarer, less well-known cancer; a lack of information and an abundance of unanswered questions."

SPOTLIGHT ON

NEUROENDOCRINE CANCER



*Long term conditions

Neuroendocrine cancer sits at the interface of rare/uncommon diseases, cancer, and, for many, chronic diseases.

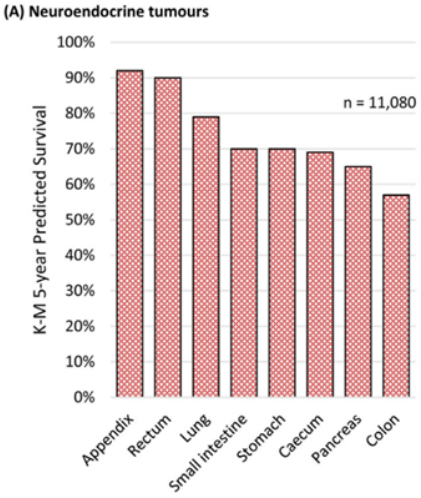
DEFINITION OF NEUROENDOCRINE CANCER

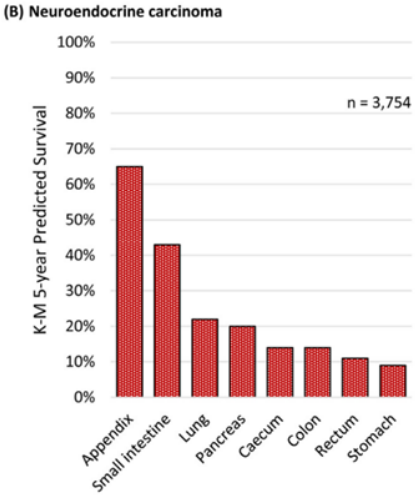
Neuroendocrine neoplasm (NEN) is the medical umbrella term for neuroendocrine cancers. This group of cancers has two WHO clearly defined classifications:

Neuroendocrine Tumours (NETs) and
Neuroendocrine Carcinomas (NECs).
Both classifications are malignant.

Neuroendocrine Tumour (NET) ~70% of all NENs	Neuroendocrine Carcinoma (NEC) ~30% of all NENs
• Well-differentiated • Slow to rapid growth - Graded 1-3 • May present/develop site- specific hormone related symptoms /syndromes	• Poorly differentiated • Rapid growth - Grade 3: large or small cell morphology • May present/develop paraneoplastic syndrome

Determining whether patients have NETs or NECs is essential. The pathologic differentiation status of these cancers can significantly impact the patient's prognosis and treatment decision. (6) There are significant differences in 5-year survival of NETs and NECs.(7)





Visit Our Website
www.neuroendocrinecancer.org.uk

SPOTLIGHT ON
NEUROENDOCRINE
CANCER

PRESENTATION

CLINICAL PRESENTATION

- Neuroendocrine cancers have a higher prevalence than incidence: incidence ~ 9 per 100,0001, prevalence ~ 35 per 100,000.(8)
- Diagnosing NENs is challenging; data and patient experience reports highlight an average time to diagnosis of 3 years: (9)
  - Symptoms may mimic or be masked by more common conditions such as IBS, Asthma or Menopause.(2)
  - Routine tests may not raise suspicion or diagnosis of NENs, e.g., small bowel NENs may have negative endoscopy and normal FBC and CEA despite bowel symptoms.
  - Symptoms may be tumour and hormone excess related. However, many may be asymptomatic at early stages, found incidentally through screening or other tests.
- Those with functioning tumours have specific symptoms. For example, those with carcinoid syndrome may present with the classical triad of diarrhoea, flushing and wheezing.

- CT +/- MRI is recommended in patients presenting with persistent abdominal pain and diarrhoea. Contrast-enhanced CT is considered the basic imaging for NC diagnosis, staging, and surveillance/monitoring, while MRI is preferred for examining the liver, pancreas and bone. Both have limitations in identifying all hyper or hypovascular NC tumour deposits. (10,11)
- Whole-body Nuclear Medicine (SSTR) imaging is recommended for neuroendocrine cancer staging/diagnosis (68Ga DOTATATE PET/CT is preferred; if not available, Octreoscan can be undertaken but may underrepresent the true extent of the disease). 18FDG-PET/CT is better suited for high-grade NENs (G2-3NETs/G3 NECs).
- Refer patients with a confirmed or highly suspected diagnosis of NET/NEC to your local NET MDT or Centre of Excellence.

The Neuroendocrine Cancer UK Stakeholder Group recommended "Neuroendocrine Cancer Pathway," launched in June 2023. Visit www.neuroendocrinecancer.org.uk for more information and the latest updates.

Neuroendocrine Cancer UK
Incorporating The Ann Edgar Trust

NEUROENDOCRINE CANCER

An overview for Healthcare professionals

UKI NETS

For clinical information and advice about NETs and NECs, including expert guidelines, please visit the Clinical Practice page on the UK and Ireland Neuroendocrine Tumour Society's website:
www.ukinets.org

Neuroendocrine Cancer UK
Incorporating The Ann Edgar Trust

If you have a patient with (NET or NEC), please signpost them to Neuroendocrine Cancer UK, a charity dedicated to providing support, advocacy, information and education to anyone affected by neuroendocrine cancer.

Office: 01926 883487 | Helpline: 0800 434 6476
www.neuroendocrinecancer.org.uk

1. White et al 2022;23: 100510 The Lancet Regional Health – Europe. Incidence and survival of neuroendocrine neoplasia in England 1995−2018: A retrospective, population- based study
2. Basuroy et al BMC Cancer 2018;18(1):1122. Delays and routes to diagnosis of neuroendocrine tumours.
3. Basuroy et al. Neuroendocrinology 2018; 107:42-9. Presenting Symptoms and delay in diagnosis of gastrointestinal and pancreatic neuroendocrine tumours.
4. Bouvier & Jervis. Current Opinion in Endocrine and Metabolic Research 2021; 18: 254-257. Patient perspectives, from diagnosis through treatments and beyond.
5. Neuroendocrine Cancer UK/RareMinds Open Conversations Report- https://www.neuroendocrinecancer.org.uk/wp-content/uploads/2022/07/Neuroendocrine Cancer UK-rareminds-Open-Conversations-report-2022_final-3.pdf
6. BSG Guidelines 2021 - https://www.bsg.org.uk/clinical-resource/british-society-of-gastroenterology-guidelines-on-the-management-of-irritable-bowel-syndrome/